OPTIMIZING THE QUALITY OF LIFE OF INDIVIDUALS
WITH HUNTINGTON DISEASE

by

Randy Goossen
Bachelor of Social Work, University of the Fraser Valley 2000

MAJOR PAPER SUBMITTED IN PARTIAL
FULFILLMENT OF
THE REQUIREMENTS FOR THE DEGREE OF

MASTER OF SOCIAL WORK

In the
School of Social Work and Human Services

© Randy Goossen 2014

UNIVERSITY OF THE FRASER VALLEY

Spring 2014

All rights reserved. This work may not be
reproduced in whole or in part, by photocopy
or other means, without permission of the author.
Approval

<table>
<thead>
<tr>
<th>Name</th>
<th>Randy Goossen</th>
</tr>
</thead>
<tbody>
<tr>
<td>Degree</td>
<td>Master of Social Work</td>
</tr>
<tr>
<td>Title of Thesis</td>
<td>Optimizing the quality of life of individuals with Huntington Disease</td>
</tr>
<tr>
<td>Examining Committee:</td>
<td></td>
</tr>
<tr>
<td>Chair</td>
<td>John Hogg, BA, BSW, MSW, RSW</td>
</tr>
<tr>
<td></td>
<td>Faculty, School of Social Work and Human Services</td>
</tr>
<tr>
<td></td>
<td>Leah Douglas, BSW, MSW, PhD, RSW</td>
</tr>
<tr>
<td></td>
<td>Senior Supervisor</td>
</tr>
<tr>
<td></td>
<td>Faculty, School of Social Work and Human Services</td>
</tr>
<tr>
<td></td>
<td>Lisa Moy, BA, BSW, MSW, PhD, RSW</td>
</tr>
<tr>
<td></td>
<td>Supervisor</td>
</tr>
<tr>
<td></td>
<td>Faculty, School of Social Work and Human Services</td>
</tr>
</tbody>
</table>

Date Defended/Approved: April 11, 2014
The author, whose copyright is declared on the title page of this work, has granted to the University of the Fraser Valley the right to lend this major paper, thesis, project or extended essay to users of the University of the Fraser Valley Library, and to make partial or single copies only for such users or in response to a request from the library of any other university, or other educational institution, on its own behalf or for one of its users.

The author has further granted permission to the University of the Fraser Valley to keep or make a digital copy for use in its circulating collection, and, without changing the content, to translate the major paper/thesis/project or extended essay, if technically possible, to any medium or format for the purpose or preservation of the digital work.

The author has further agreed that permission for multiple copying of this work for scholarly purposes may be granted by either the author or the Associate Vice-President, Research and Graduate Studies.

It is understood that copying or publication of this work for financial gain shall not be allowed without the author’s written permission.

Permission for public performance, or limited permission for private scholarly use, of any multimedia materials forming part of this work, may have been granted by the author. This information may be found on the separately catalogued multimedia material and in the signed Partial Copyright License.

The original Partial Copyright License attesting to these terms, and signed by this author, may be found in the original bound copy of this work, retained by the University of the Fraser Valley Heritage Collection.

University of the Fraser Valley
Abbotsford, B.C.
I hereby grant the University of the Fraser Valley the right to lend my major paper / thesis / project / extended essay to users of the University of the Fraser Valley Library or to other libraries. Furthermore, I grant the University of the Fraser Valley Library the right to make single copies only of my major paper / thesis / project / extended essay for users of the library or in response to a request from other libraries, on their behalf or for one of their users.

I further grant permission to the University of the Fraser Valley to keep or make a digital copy for use in its library collection and / or institutional repository.

Permission for extensive copying of this major paper / thesis / project / extended essay may be granted by me or by a member of the University designated by me.

It is understood that copying or publication of this major paper / thesis / project / extended essay for financial gain shall not be allowed without my written permission.

Permission for public performance, or limited permission for private scholarly use, or any multimedia materials forming part of this work, is covered by a separate agreement if applicable.

Title of major paper / thesis / project / extended essay:

**Optimizing the quality of life of individuals with Huntington Disease**

Author (print name): **Randy Goossen**

Signature: ____________________________________________

Date: ____________________________________________
Abstract

Huntington Disease is a genetically inherited neurological disorder that most often strikes adults in mid-life (between 30 and 55 years of age). Each child of a parent with Huntington Disease is at 50% risk of inheriting the disease themselves. While genetic testing has been available to determine the presence of the mutant gene since 1993, those at risk struggle with the decision whether or not to be tested as there is no cure or treatment capable of stopping the progression of the disease.

The purpose of this paper is to examine how some individuals who have Huntington Disease are able to live lives of high quality while others, similarly affected by the disease, spiral downward emotionally, cognitively and physically with some even taking their own lives. The researcher sought to identify themes, characteristics, supports, services and resources, as well as examine the common variables amongst those who were thriving despite living with ever-advancing Huntington Disease.

The exploratory research was conducted through the use of focused interviews with Huntington Society of Canada professional staff from across Canada who work with individuals with Huntington Disease and their families.

Findings from this research suggest social determinants of health, clients’ degree of self-awareness, family history with the disease, stigma associated with the disease and the availability of supports and services in their area all contribute to clients’ well-being. Research participants suggested clients’ social connectedness and their own attitudes towards the disease, their personalities and their receptiveness to support and assistance are likely the greatest factors impacting clients’ quality of life.
Acknowledgements

I would like to acknowledge the Huntington Society of Canada for their support and encouragement in conducting my research as well as the research participants for their time, wisdom and for the generous contributions they shared which formed the basis for this project. Special thanks also go to Susan Tolley and Dr. Jessica Easton who have been incredible mentors and supports to me through the years, particularly early on when I was first introduced to the Huntington Disease community. I would also like to extend my gratitude and respect to the individuals and families who live with Huntington Disease and who demonstrate incredible grace, dignity and resiliency as they travel their journeys with the disease sharing their struggles, their stories and their lives with the Family Services Team of the Huntington Society of Canada.

I would also like to thank Dr. Leah Douglas, Dr. Lisa Moy and the rest of the Social Work faculty at the University of the Fraser Valley for their instruction, support, encouragement and wisdom through the completion of this research.

I also want to thank my kids, Emma and Dylan for their patience, understanding and their willingness to share their dad over the years as I delved deeper and deeper into the fascinating and challenging reality of life with Huntington Disease.

Finally, I would like to thank Ed who first introduced me to Huntington Disease and without whom my own journey into Huntington Disease and my subsequent research would have never taken place; this research and this paper is dedicated to you Ed. This research exists because of your strength and spirit.

I sincerely hope and believe through working together, we will soon be able to ensure future generations will be free from the challenges and struggles created by Huntington Disease.
# Table of Contents

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abstract</td>
<td>v</td>
</tr>
<tr>
<td>Acknowledgements</td>
<td>vi</td>
</tr>
<tr>
<td>Table of Contents</td>
<td>vii</td>
</tr>
<tr>
<td>List of Acronyms</td>
<td>ix</td>
</tr>
<tr>
<td>List of Tables</td>
<td>ix</td>
</tr>
<tr>
<td>Introduction</td>
<td>1</td>
</tr>
<tr>
<td>Background and Significance of Problem</td>
<td>2</td>
</tr>
<tr>
<td>Literature Review</td>
<td>9</td>
</tr>
<tr>
<td>Caregivers</td>
<td>10</td>
</tr>
<tr>
<td>Stigma and Isolation</td>
<td>14</td>
</tr>
<tr>
<td>Suicide</td>
<td>15</td>
</tr>
<tr>
<td>Proactive Strategies to Improve Quality of Life</td>
<td>17</td>
</tr>
<tr>
<td>Gaps in the Research – Moving Forward</td>
<td>19</td>
</tr>
<tr>
<td>Theoretical Framework</td>
<td>20</td>
</tr>
<tr>
<td>Design and Methodology</td>
<td>22</td>
</tr>
<tr>
<td>Ethical Considerations</td>
<td>24</td>
</tr>
<tr>
<td>Study Limitations</td>
<td>26</td>
</tr>
<tr>
<td>Findings</td>
<td>27</td>
</tr>
<tr>
<td>Defining Quality of Life</td>
<td>28</td>
</tr>
<tr>
<td>Self-Awareness</td>
<td>29</td>
</tr>
</tbody>
</table>
Stigma 31
“Self” and Huntington Disease 34
Family and Huntington Disease 36
Community and Geographical Variability 37
A Little Help from my Friends 39
Grief and Loss 40
Advance Care Planning 41
Consumer Directed Care 43

Implications for Practice, Policy and Research 44

Dissemination 46

Recommendations for Future Research 47

Conclusion 48

References 50

Appendices

Appendix A: Informal E-Mail Advising of Research 56
Appendix B: UFV Ethic Board Certificate 57
Appendix C: Recruitment E-Mail 58
Appendix D: Letter of Consent 59
Appendix E: Participant Interview Guide 63
### List of Acronyms

<table>
<thead>
<tr>
<th>Acronym</th>
<th>Meaning</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACP</td>
<td>Advance Care Planning</td>
</tr>
<tr>
<td>BC</td>
<td>British Columbia</td>
</tr>
<tr>
<td>FST</td>
<td>Family Service Team</td>
</tr>
<tr>
<td>FSW</td>
<td>Family Service Worker</td>
</tr>
<tr>
<td>HD</td>
<td>Huntington Disease</td>
</tr>
<tr>
<td>HDRC</td>
<td>Huntington Disease Resource Center</td>
</tr>
<tr>
<td>HSC</td>
<td>Huntington Society of Canada</td>
</tr>
<tr>
<td>MSW</td>
<td>Master of Social Work</td>
</tr>
<tr>
<td>RCD</td>
<td>Resource Center Director</td>
</tr>
<tr>
<td>SW</td>
<td>Social Work</td>
</tr>
<tr>
<td>UFV</td>
<td>University of the Fraser Valley</td>
</tr>
</tbody>
</table>

### List of Tables

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Shoulson-Fahn Functional Capacity Rating Scale</th>
<th>pg. 5</th>
</tr>
</thead>
</table>
Introduction

What gives life meaning, purpose, and quality? How is an individual’s quality of life impacted if diagnosed with a progressive neurological disorder whose course would slowly rob the individual of their independence? How receptive might one be to consider available supports and services if part of the disease’s progression involved losing the ability to assess needs or plan and anticipate pending losses? How would one’s quality of life be affected if family roles were thrown into disarray? How might one accept their children prematurely becoming caregivers, or what would one do if they found themselves thrust in a downward spiral reminiscent of a spiral they may have witnessed or supported an affected sibling or parent descend?

These questions are important as they are the reality for individuals who carry a diagnosis of being gene positive for Huntington Disease (HD). While a great deal has been written about quality of life and HD, little research has been done on how individuals with HD perceive their own quality of life. Research is also scarce on identifying the factors that contribute to how some individuals thrive despite the disease progression, while others who may be similarly affected, appear to steadily deteriorate.

The purpose of this research is to explore the factors that contribute to the quality of life for individuals with HD and to contribute to social work practice knowledge about this topic. The research will inform how services, supports, resources and information can be adapted, promoted, introduced and delivered to enhance and improve the quality of life of those individuals impacted by HD. This research will identify common themes and factors which allow some individuals with HD to optimize the quality of their lives and will identify the most critical ingredients to assist others with HD do the same.
The research conducted was inspired by my own observations and experiences as a contracted employee with the Huntington Society of Canada (HSC) over the past nine years; through my own practice experience, I have had the opportunity to observe clients’ incredible strength of spirit and resiliency as well as their ability to continually adjust and adapt to the disease’s ongoing progression. Through volunteering at the annual British Columbia Therapeutic Retreat for individuals with HD, I have had the opportunity to work with dozens of individuals with HD and had become increasingly curious what those who were thriving despite having HD shared in common. While it was initially hoped the research study could be conducted directly with individuals with HD, it became evident this would be impractical due to time constraints around gaining research ethics approval. As a result, the research was conducted with professional staff employed by the HSC who work directly with clients and families affected by HD.

**Background and Significance of Problem**

*Huntington Disease* (HD) can be defined as a genetically inherited progressive neurodegenerative disorder which affects patients’ cognitive, emotional and motor functions. HD causes severe disability, often becoming evident when individuals are in the prime of their lives (Banaszkiewicz, Sitek, Rudzinska, Soltan, Slawek, & Szczudlik, 2012). The onset of HD can be gradual and progressive, or rapid and severe. HD can include symptoms such as unsteady gait, clumsiness, involuntary movements (chorea), slurred speech, and swallowing difficulties and can also cause psychiatric symptoms such as depression, apathy, mood swings, personality changes, and impaired judgment (Roscoe, Corsentino, Watkins, McCall, & Sanchez-Ramos, 2009). Paulsen (as cited in Halpin, 2012) suggests “the disease is a genetic, degenerative and fatal condition with symptoms including mood imbalances, memory loss, personality changes
and chorea” (p.318). Pogledic and Relja (2012) report HD was discovered and was first described by Dr. George Huntington in 1872 and was originally called Huntington’s Chorea, chorea for the dance like movements that frequently accompany the disease. Susan Tolley, British Columbia Huntington Disease Resource Center Director (BC-HDRC) (personal communication, 2005) advised since it has been discovered chorea is not always a part of the disease process, the word chorea has largely been eliminated when describing HD, although the term chorea is still used occasionally to specifically describe the involuntary movements.

HD is a disease with identifiable biomarkers, making diagnosis possible prior to the presence of symptoms. A positive diagnosis for HD can be confirmed while individuals are presymptomatic, while symptoms are mild, or a diagnosis may be made long after symptoms have become severe. While some individuals at risk of carrying the mutant HD gene due to family lineage choose to go through the genetic testing process, many others choose not to be tested and leave their fate and the fate of their children unknown. Babul et al. (1993) report attitudes towards direct predictive testing for the HD gene vary with data on the actual rate of testing suggesting the rate is much lower than predicted by attitudinal surveys. The percentage of people at risk for HD who requested testing when approached by registries or testing centers varied from 9% in Wales, 10% in Indiana, and 16% in the Manchester area of England. According to genetic counsellor Susan Creighton (personal communication February 11, 2014) at the HDRC at the University of British Columbia Hospital in Vancouver, British Columbia, 80% of those at risk of inheriting the mutant HD gene choose not to have genetic testing done.

HD impacts the whole person and while the motor, cognitive and behavioral disturbances associated with HD are well documented, Ho and Hocaoglu (2011) note “there is little empirical data examining how this translates into what patients themselves are concerned about throughout
the long course of the disease” (p.235). Similarly, there is limited research on how the presence of the disease impacts the quality of individuals’ lives. Perry (1981) advises, “while some patients manage to go through the entire illness without a great deal of suffering, for most it is a devastating experience” (p.1098). Ultimately though, how individuals and their families define quality of life and how they define HD may provide the greatest indication of how individuals will carry themselves and their families through the disease process. As Carlozzi and Tulsky (2012) report a participant in their study suggested “it’s not how you’re going to die… it’s how you’re living or how you’re gonna live” (p.223) that best determines an individual’s quality of life.

HD presents and manifests itself in a variety of ways and varies greatly from individual to individual. Ho and Hocaoglu (2011) suggest monitoring and charting the course of the disease is important as it provides professionals and caregivers the opportunity to be aware of the phases and changes through the disease progression and provide “an informed basis for the long-term management of health and well-being in HD, and the development of interventions across the spectrum of disease phases” (p.238). In order to provide a common language when discussing HD progression, Shoulson-Fahn developed the following Functional Capacity Rating Scale in 1979 which still remains a widely used reference when assessing the stage of HD progression with clients:
<table>
<thead>
<tr>
<th>Stage</th>
<th>Usual level</th>
<th>Lower level</th>
<th>Marginal</th>
<th>Unable</th>
<th>Unable</th>
<th>Unable</th>
<th>Unable</th>
<th>Home</th>
<th>Home</th>
<th>Home</th>
<th>Home or extended care facility</th>
<th>Total care facility only</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Full</td>
<td>Requires slight assistance</td>
<td>Requires major assistance</td>
<td>Unable</td>
<td>Unable</td>
<td>Unable</td>
<td>Severely impaired</td>
<td>Care can be provided at</td>
<td>Home</td>
<td>Home</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Full</td>
<td>Full</td>
<td>Impaired</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Full</td>
<td>Full</td>
<td>Mildly impaired</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td>Home</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Full</td>
<td>Full</td>
<td>Unable</td>
<td>Home or extended care facility</td>
<td>Home</td>
<td>Home</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Full</td>
<td>Full</td>
<td>Unable</td>
<td>Total care facility only</td>
<td>Home</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

This table was adapted from Shoulson and Fahn, 1979 from: http://www.stanford.edu/group/hopes/cgi-bin/wordpress/2010/07/assessment-standards-for-huntingtons-disease-severity/

Those who are living at risk of HD struggle with determining what it means to live at risk. Many individuals at risk live for years, fearing and often assuming they are gene positive. Other individuals who have been tested and who have confirmed they are gene positive, acknowledge the reality of having the disease, yet still manage to live full, complete and active lives despite their diagnosis and despite knowing the disease progression is sure to follow. While there is no standard response to learning of the presence of HD for an individual or a family, there is no doubt, HD itself along with its myriad of presentations will surely impact the lives of those affected.

Deciding whether or not to be tested for the mutant HD gene is a deeply personal matter with far reaching implications. Despite the availability of genetic testing, which is able to conclusively determine whether or not an individual carries the gene, only a small percentage of those at risk of developing HD choose to have it prior to the development of visible symptoms (Andersson, Juth, Petersen, Graff & Edburg, 2012). It appears the vast majority of those at risk choose to live with the unanswered question of whether or not they will develop the disease.
Many struggle with the awareness that in the event they choose to have children, the children will also inherit a 50% chance of carrying the mutant gene (if the parent turns out to be gene positive). Each individual at risk for HD is faced with a decision to test for HD. They must decide if they will have a higher quality of life from the knowledge that they will acquire the fatal disease or if they will have a higher quality of life if their genetic fate is left unknown.

The term *quality of life* itself is subjective, in that one person’s pleasure can be another’s pain. In an effort to examine and ultimately optimize the quality of the lives of individuals impacted by HD, it is important to define quality of life when examining the literature. The World Health Organization (1997) defines quality of life as “the individual’s perception of the patient’s position in life, in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards, and concerns” (p.1). Carlozzi and Tulsky (2012) differentiate health related quality of life as “a multidimensional construct” which they suggest differs from a more generalized definition of quality of life which they describe as “a less clearly defined one-dimensional construct that evaluates general well-being or life satisfaction” (p.213).

For the purpose of the research being conducted, the definition from the World Health Organization will be applied as this definition provides a holistic, person-centered and potentially strength-based definition which is in alignment with both the researcher’s own values and ethics as well as with the values and ethics of social work (SW) as a profession.

While one’s quality of life is ultimately subjectively defined by each individual, broader structural factors beyond the control of individuals also play a significant role in determining quality of life as well as what options might be available to individuals. According to Raphael (2007) social determinants of health can be defined as “the economic and social conditions that shape the health of individuals, communities, and jurisdictions as a whole” (p.19). Raphael
(2007) further suggests social determinants of health are comprised of “aboriginal status, early life, education, employment and working conditions, food security, health services, housing, income and income distribution, social exclusion, social safety net and employment and unemployment insecurity” (p.19). Individuals impacted by HD also face structural barriers and are often hard-pressed to access resources and supports that are much more easily accessed by others.

The Huntington Society of Canada (HSC) has played a significant role in supporting generations of individuals and families affected by the disease since the society was founded in 1973 (Huntington Society of Canada, 2014). The HSC acknowledges the diversity and variability of services across Canada and strives to provide a common thread and play an important role nationally, provincially and regionally in promoting the well-being of individuals and families affected by HD. The HSC is also key stakeholder and an active participating agency in integrating and collaborating on global research efforts aimed at identifying effective treatments and eventually finding a cure for HD. The HSC is also committed to supporting efforts to maximize the quality of life of individuals and families living with, or at risk for HD, as is evident in the HSC Mission Statement (HSC Website):

The Huntington Society of Canada aspires to a world free from Huntington disease. The Society maximizes the quality of life of people living with HD by:

Delivering services;

Enabling others to understand the disease and:

Further research to slow and prevent Huntington disease.
The HSC also provides a gateway for individuals wishing to become involved in clinical trials and invites and encourages participation from those with HD who want to be a part of finding answers to the many unanswered questions regarding HD.

While the HSC focuses its efforts on supporting research initiatives, they also provide resources aimed at supporting and improving the quality of life of those affected by the disease. The primary means of delivering these supports is through the HSC Family Services Team (FST). Research participants were comprised of professionals with social work backgrounds (70% of respondents), psychology backgrounds (12% of respondents) and Bachelor of Arts (18% of respondents). Social work education and training is especially well-suited to the needs of the HD community and helps ensure the HSC FST is well-equipped to provide support, education, counseling and advocacy for clients and families. HSC FST staff facilitate support groups as well as provide in-services and training for professional staff in a variety of institutional settings who provide care to individuals with HD in hospitals, care facilities, correctional facilities as well as in a variety of other settings.

Through my own work with individuals and families with HD, I have witnessed incredible resiliency, adaptability, creativity and incredible strength of spirit and determination in those confronting HD; these characteristics are certainly worthy of further research. Through my position as a Family Service Worker (FSW) with the HSC, I have also been privy to individuals and families’ struggles with HD and have witnessed first-hand the benefits of being connected to the HD community as opposed to those who choose, or who see no other option but to travel their journey with HD on their own.
It is my hope that common threads and themes can be identified within this research which will contribute to a body of knowledge to help support and encourage people with HD to adapt and adjust to the progressive changes and challenges posed by living with this devastating disease. It is also my hope social workers and other professional staff working with individuals and families impacted by HD will reflect on their own practices and will carefully consider the social positioning of those whom they serve. Guo and Tsui (2010) suggest practitioners “respond intuitively to the strengths of everyday life, and will empower disadvantaged people to discover their capacity to resist inequality and overcome adversity” (p.242).

**Literature Review**

Considerable research on Huntington disease has been amassed since the disease was first thoroughly described by George Huntington in 1872 (Halpin, 2012). Prior to 1993, diagnosis for HD was based on demonstrated symptoms and through detailed family histories when available. However, since 1993 when the specific gene which causes HD was identified, it has become possible to make an accurate diagnosis prior to at-risk individuals experiencing symptoms; since this discovery, research into HD has further intensified. Bombard et al. (2009) report HD had become the first autosomal genetic disease with a predictive test allowing those at risk to know with certainty whether they have inherited the gene mutation before developing symptoms. With the availability of genetic testing, many questions arise as to the ethics of, and the benefits of predicting a disease that does not yet have an effective treatment or cure. Andersson et al. (2012) question whether “the knowledge that you or a close relative will develop a fatal disease actually lead to a better quality of life or do the anxiety and sadness resulting from confirmation of the disease outweigh the possible benefits” (p.190). The authors explain further:
Being able to predict whether someone will develop a serious disease in the future is associated with several ethical issues, such as who is entitled to information regarding the results of a genetic test, if, how and when they should be disclosed, as well as the consequences of the result for the person taking the test and their family. (Andersson et al., 2012, p.190)

Whether or not to be tested is clearly a deeply personal question which will be discussed later in this paper.

The focus of this paper will be primarily on unpaid, informal and family caregivers, stigma, isolation, suicide as well as exploring pro-active strategies individuals and caregivers can employ to improve the quality of life of those living with HD. Existing gaps in the literature will also be identified and will be discussed within this paper.

**Caregivers**

Those who are gene positive for Huntington disease often recognize the onset of symptoms in mid-life. HD then impacts not only the individual directly affected, but also the entire family system. While caregivers might emerge from within the family and rise to the challenge of providing and/or coordinating care, others with HD are less fortunate and lack available or willing family caregivers and are forced to rely on the support of friends or other informal support networks, or they may be even more isolated and be forced to face the many challenges of living with HD on their own. Literature on quality of life and HD frequently mentions the challenging role of the caregiver. The genetic implications of HD and the potential for HD repeating itself within the family, thereby threatening successive generations, contributes to further aggravate and complicate the care-giving role (Aubeeluck, 2005). In many families,
caregivers are sons or daughters, who themselves may be at risk or who may already know they are gene positive, yet they very well could find themselves responsible for the care and well-being of their loved ones. Carlozzi and Tulsky (2013) caution of the risk of “forward comparisons” (pg.224) which can result in caregivers anticipating their own futures fearing the life they believe they may have in store for themselves. O’Connor and McCabe (2012) suggest “caring for a partner or family member with a progressive neurological illness has been recognized as being a source of burden and distress resulting in lowered levels of quality of life” (p.703). This reduced quality of life is assumed before even considering the additional genetic element of HD which further aggravates and greatly complicates the already difficult care-giving role.

The diagnosis of being gene positive for HD triggers the start of a journey down a path characterized by progressive neurological decay and continued loss of independence. Mitchell, Kemp, Benito-Leon and Reuber (2010) suggest quality of life in those with HD has been poorly studied, especially in relation to cognitive change. However, the diagnosis does not necessarily guarantee a diminished quality of life for those diagnosed, nor for caregivers. Helder, Kaptein, Van Kempen, Weinman, Van Houwelingen & Roos (2002) hypothesize that perception of illness and mechanisms for coping both contribute greatly to patients’ and caregivers’ quality of life, while Cella (1995) offers hope and suggests despite the inevitable progressive motor, cognitive and psychiatric deterioration, individuals can show some improvement in health-related quality of life over time. Kapstein et al. (2007) suggest patients and their caregivers perceive illness differently, with the perceptions themselves impacting quality of life; an individual with HD might fail to recognize a challenge obvious to a caregiver, or the person with HD may alternately perseverate over an issue the caregiver regards as an insignificant matter. O’Connor and
McCabe (2012) write that caregivers and individuals with HD also report having differing perspectives on marital satisfaction with those with HD reporting greater levels of satisfaction than do their care-giving partners. Banaszkiewicz et al. (2012) conclude the ideal way to obtain a realistic picture of the symptoms and difficulties experienced by an individual with HD is to consider the perspectives of the person with HD, family members, as well as the perspectives of the medical professionals involved. The HDRC and other multi-disciplinary assessment and treatment teams typically request the active involvement, participation and perspective of patients’ families and caregivers through the assessment and care planning process in order to obtain a more accurate perspective on how the individual with HD is truly managing.

While it stands to reason that the presence of a caregiver in the life of a person with HD would be of benefit to the person with HD, the perception the caregiver has of their role and relationship with the person with HD further contributes to the quality of life of the person for whom they are providing care (Kaptein et al., 2007). Despite the inherent challenges of providing care to another, caregivers must also be mindful of the impact their negative or critical perception might have on their loved one. Helder et al. (2002) acknowledge the challenge of being a caregiver, suggesting “spouses are often the forgotten members of the family with HD” (p.39), yet are relied upon to maintain all of the responsibilities of the home as well as for their own feelings regarding their role. Roscoe et al. (2009) suggest little is known about the general well-being of caregivers for individuals with HD and suggest there is much progress to be made mitigating the long-term stresses of care-giving. Research however, appears to reveal literature is much more plentiful on the quality of life of caregivers than it is on examining the quality of life of those individuals who are living with HD themselves.
Research suggests chronically ill individuals and their caregivers perceive both illness and quality of life very differently with caregivers typically reporting greater symptoms and a lower quality of life for their loved one, than does the individual directly affected by the chronic disease themselves (Kapetein et al., 2007). Hocaoglu, Gaffan and Ho (2011) suggest patient-reported quality of life assessments are subjective; therefore, it is important to examine the caregivers’ ratings of patients’ quality of life, especially in light of the cognitive impairment associated with HD. A great deal of research has been done on quality of life and many measures and scales have been developed to assess quality of life (Burckhardt and Anderson, 2003; Hocaoglu et al., 2012 & Carlozzi and Tulsky, 2012). However, according to Carlozzi and Tulsky (2012), far less research and far fewer measures have been created to measure health-related quality of life. Hocaoglu et al. (2012) have endeavored to address this need through the development of an HD specific health-related quality of life questionnaire. Although the development of a HD specific assessment tool has been a valuable contribution in assessing and measuring the quality of life of individuals with HD, the development and application of a standardized and widely accepted HD specific quality of life measurement tool remains in its infancy and still requires a great deal of research and development.

With caregivers playing a critical role in the overall well-being and quality of life of individuals with HD, it is imperative the caregivers’ well-being is also supported throughout the disease progression. Caregiver burden has been a long-standing challenge for those providing care, with Perry (1981) suggesting “if spouses and other family members were given practical help in caring for patients, as well as regular planned holiday relief, a greater proportion of patients could have the comfort of living at home through most of their illness” (p.1100). Caregivers as well would reap benefits as they could take much needed time away from their
care-giving duties and could thereby sustain both themselves and those to whom they are providing care.

Stigma and Isolation

Huntington disease strikes individuals on multiple levels affecting movement, emotion and cognition. While symptoms vary between individuals, frequently the involuntary movements, speech problems, unsteady gait and intoxicated appearance commonly associated with HD, lead to the stigmatization and isolation of both the individual with HD and their caregivers (Helder et al., 2002). Aubeeluck and Buchanan (2007) suggest the mood, behavior and movement disorders associated with HD often lead to caregivers and individuals with HD isolating themselves from social outings and can contribute to chronic isolation for both the HD and non-HD affected spouse. Hayden et al. (1980) report HD related anti-social behavior may also contribute to caregivers’ fears of social embarrassment and of possible rejection by friends. O’Connor and McCabe’s (2011) research indicates that following a neurological diagnosis, caregivers reported “the number of friends dropping by for a visit, inviting the carer out socially … reduces dramatically and was often noted as learning who their real friends are” (p.704). Many individuals and families impacted by HD avoid social situations entirely often citing secrecy around the disease and the lack of awareness and understanding others had about HD as their reasons; for example as Carlozzi and Tulsky (2012) report a participant in their research reported “a lot of people think I’m drunk or on drugs” (p.219). When the stigma and lack of understanding around HD is combined with problems initiating, difficulty with motor tasks and frequently depression, it is not surprising those with HD often find maintaining social contacts and community activities more trouble than they are prepared for, which of course then leads to even further chronic isolation.
Stigma and isolation are often present both within and beyond the social arena. Perry (1981) reported in many jurisdictions insurance is denied, licenses are revoked and HD affected applicants are refused employment as a result of existing symptoms as well as in anticipation of worsening symptoms in the future. Dellefield and Ferrini (2011) caution that the disease progression which can include bizarre and impulsive behavior, psychotic symptoms, poor judgment and an intoxicated appearance can lead to the involvement of law enforcement, mental health and can eventually lead to institutionalization.

The stigma and isolation associated with HD clearly takes a considerable toll with Engstrom and Nordeson (1995) warning “to be able to live with others gives you strength and courage to face life, it is dangerous to live in isolation” (p.182). Unfortunately though, isolation becomes a reality for all too many individuals living with and progressing through the advancing stages of HD and this isolation can lead individuals with HD to take desperate, and sometimes irreversible, actions.

Suicide

HD can contribute to progressive loss of neurological function, motor control, loss of cognitive skills as well as changes in mood and behavior (Williams, Skirton, Barnette, & Paulsen, 2011). A lack of self-awareness of their own symptoms (Carlozzi and Tulsky, 2012) and the accompanying risks as well as the inability to resolve complex and multi-dimensional challenges are also often part of the HD disease process. Halpin (2012) reports Dr. George Huntington believed HD was akin to insanity leading to suicide and Halpin further reported suicide to be the second to fifth leading cause of death amongst those with HD. It is no wonder that there are high rates of suicide given that so many individuals with HD struggle with
loneliness, isolation, depression and desperation. Far too often, when faced with what might appear to be insurmountable barriers and a bleak future, a disproportionately high number of individuals with HD choose suicide as an end to their problems.

In his research on suicide within the HD community, Halpin (2012) discovered suicide to be a major cause of death with an American study suggesting 9.3 – 13% of all deaths in the HD community being suicides, in comparison to 1 to 1.5% in the general population. Other American studies on suicide amongst the HD population estimate the suicide rate at 3 to 23 times that of the general population (Roos, 2010; Schoenfeld et al., 1984). Coustasse, Pekar, Sikula and Lurie (2009) point to the challenges of making decisions around genetic testing, as well as the ethical implications, such as potentially passing the gene mutation on to future generations, as potentially aggravating factors for suicide, while Paulsen, Hoth, Nehl and Stierman (2005) argue genetic testing may actually serve to reduce suicide risk, suggesting suicide rates appear to be higher in symptomatic but undiagnosed individuals when compared to those who have undergone genetic testing.

With suicide, depression, and apathy so pervasive amongst people with HD, Perry (1981) suggests it is important the isolation surrounding the disease be reduced to lessen the suffering of both individuals with HD and their caregivers. Halpin (2012) suggests in his research “both caregivers and individuals with HD expressed deep concern over what they perceived would be a slow, drawn out death from the disease” (p.330). It is therefore critical that efforts be made to reduce isolation, instill hope and contribute to factors and strategies to improve the quality of life of those with HD as well to help reduce the high rates of suicide amongst the HD community. Travers, Jones and Nicol (2007) suggest:
Huntington’s disease is a chronic neurodegenerative condition and there is evidence to suggest that clients and families affected by HD may be in need of palliative care as the disease progresses. It is also a condition with a wide range of presenting and ongoing physical and psychological symptoms which potentially need to be supported by the full, multi-professional health and social care team. Accordingly, all staff working with people affected by HD should have knowledge of the advanced stages of this condition and an understanding of the advantages of applying a palliative care approach to their care at all stages of the illness” (p.130).

**Proactive Strategies to Improve Quality of Life**

While there are limited proactive strategies offered within existing literature on improving the quality of life of those living with HD, there are several approaches that have been identified which have been reported as having met with some success. Dellefield and Ferrini (2011) offer several suggestions for managing the advanced stages of HD until death in the areas of social interactions, communication, safety, spirituality, entertainment and nutrition and functional competence. Specifics around Dellefield and Ferrini’s (2011) suggestions to improve quality of life include: making deliberate efforts to involve those with HD in daily activities; paying attention to personal choices and preferences especially as communication difficulties present increased challenges; remaining aware of physical, emotional and psychological comfort; attempting to maintain safety and order while also respecting rights to autonomy; remaining mindful of spiritual needs and the need for meaningful connections with others; coordinating activities and outings appropriate to individual interests and abilities; maintaining nourishment and nutrition while also respecting food preferences; and finally, encouraging and promoting independence as much as is possible. Having both professional paid, formal and informal
caregivers aware of proactive strategies to enhance quality of life is especially important as Traverse, Jones and Nicol (2007) report people with HD and other non-malignant diseases often experience extended palliative phases and can suffer diminished quality of life for an extended period of time. While many of the ideas offered by Dellefield and Ferrini (2011) are intended for those in the very late stages of the disease, several of the suggested offerings could be adapted and might be beneficial to individuals at earlier stages of the disease process as well.

Meaningful involvement in activities such as support groups, volunteering and visiting with friends and the maintenance of social contacts can contribute to individuals’ overall sense of well-being (Carlozzi and Tulsky, 2012). Maslow (1970) as cited in Engstrom and Nordeson (1995), stresses “social contacts and relations are profoundly human needs” (p.182). O’Connor and McCabe (2012) suggest primary relationships ought to be nourished and preserved through deliberate and mindful efforts, further adding, those involved in intimate relationships have reported they sustain stronger relationships and a higher quality of life if they have attended counseling together to maintain their relationship through the course of the disease. Gutierrez et al. in Guo and Tsui (2013) suggest “empowerment involves helping powerless people to recover their potential and strength, thereby allowing them to overcome their difficulties in everyday life” (p.235). Social workers and professional staff working with individuals with HD would be well advised to remember, while HD is progressive degenerative disorder, clients can and do develop new skills and new means of coping and despite losing physical strength, the power of their strength of spirit should not be underestimated.

Strength of spirit is difficult to define but, for the purpose of this paper, could be characterized by hopefulness, optimism and resiliency as well as through a willingness to adjust and adapt to life’s challenges and changes. Kralik, van Loon and Visentin (2006) suggest
“connecting with others and sharing experiences and stories was central to strengthening resilience among people with chronic illness” (p.18). While O’Connor and McCabe (2012) report those who are able to make effective adjustments are more likely to draw on social supports to provide them with resources and emotional support through the course of their disease progression. Engstrom and Nordeson (1995) suggest “factors like ‘fighting’ spirit are important, i.e. at least fighting against getting worse and trying to go on living with dignity and self-respect” (p.181). Each person living with HD is on their own journey and is equipped uniquely to address the challenges they face. Social workers and other professional staff working with those with HD would do well to remember to build on each client’s unique strengths and to strive to empower disadvantaged people to expand their capacity to resist inequality and to overcome adversity (Guo & Tsui, 2010).

**Gaps in the Research – Moving Forward**

While numerous studies exist examining the quality of life of caregivers, (Helder et al., 2002; Roscoe et al. 2009; Banaszkiewicz et al., 2012; Aubeeluck & Buchanan, 2007; Aubeeluk, 2005; O’Connor & McCabe, 2012) a great deal of work is yet to be done to better assess the self-rated quality of life of people living with HD. Hocaoglu et al. (2011) suggest the HD Health Related Quality of Life Questionnaire aimed at capturing the “true impact of living with the disease” (p.117) is a great starting point in assessing quality of life. However, the researchers acknowledge the HD Health Related Quality of Life Questionnaire needs to carefully consider the cognitive impairment experienced by many individuals with HD and suggest it is important participants’ self-reports are examined along with the input from caregivers whenever possible in order to gain an accurate picture of how the individual with HD is truly managing. It also appears little research has been conducted to examine the variance in quality of life of those
individuals with HD who have the active involvement of a spouse or caregiver, when compared with those who have to progress through the disease process without familial support. With HD being a disease that impacts families, it is critical supports and services be designed and targeted to the needs of those diagnosed with HD, those who are at risk, as well as those who are caregivers. O’Connor and McCabe (2010) suggest services aimed at both those living with neurological illnesses and their caregivers must acknowledge the need to maintain social activities and contacts beyond the caregiver and care recipient relationship. Existing research leaves much opportunity for further investigation into identifying what proactive steps and strategies individuals with HD and their caregivers can utilize to maintain, support and optimize the quality of life of those living with HD.

**Theoretical Framework**

The research conducted was approached from a strength based and client centered perspective (Egan, 1998). This approach is in stark contrast to the pathology-focused medical model that tends to maintain a primary focus on deficits and the negative symptoms that result from disease process. For the purpose of this research, strength based and client centered perspectives were chosen for use primarily due to the compatibility with the researcher’s own practice experiences with individuals living with HD who have demonstrated the ability to find creative and unique ways to thrive despite the debilitating effects of the disease. A strength based approach focuses on clients’ self-determination and draws from clients’ previous successes in facing and addressing previous challenges and struggles. A client centered approach begins where clients’ are and focuses on identifying, defining and endeavoring to reach goals and objectives deemed to be important or desirable to clients (Egan, 1998). This positive and holistic approach was also chosen due to its “fit” between the researcher’s own values, practice
philosophy, and the material being researched. Each client’s own previous life experiences, their successes and their failures facing challenges in the past, as well as the existence of a support system and/or a willingness to create a community of support, all contribute to how individuals will ultimately approach their own unique journey with HD.

Norman (as cited in Guo & Tsui, 2010) suggests “our strengths make us resilient in periods of adversity, so social workers must cultivate and enhance the strengths of their service users” (p.234). The researcher determined it was critical the research be approached from a client-centered perspective as the disease impacts each individual and family system differently and each individual’s and family’s coping style, strengths, resources, assets and needs are unique to each family’s circumstances. It is critical social workers working with clients and families with HD identify, bolster, enhance and encourage the further development of existing strengths and assets within the client’s system of support by identifying, celebrating and building upon previous successes. Social workers ought to also support clients and families to recognize and repeat those things that are working, while also identifying and exploring possible options in areas where greater improvement is desired or could be realized. If clients are very isolated and are largely facing HD alone, social workers and other professionals working with the client must identify clients’ own potentially untapped assets as well as create bridges and linkages between clients and both the informal and formal services and supports available.

While strength based and client centered approaches can be motivating, inspiring and empowering for clients who are both challenged and supported to reach their full potential in overcoming and addressing barriers and challenges, very real and seemingly immovable barriers may not be addressed as effectively by such individual focused approaches. Structural barriers such as the impacts of social determinants of health and systemic challenges also significantly
impact individuals’ ability to address the challenges they face. Socially connected, educated, privileged, and insurance covered individuals are all likely to manage more successfully than their less connected and less privileged peers.

**Design and Methodology**

The research conducted was an exploratory study, and invited participation from key informants from the Huntington Society of Canada (HSC). Given the relatively small number of Resource Center Directors (RCDs) and Family Service Workers (FSWs) employed by the HSC across Canada, invitations for participation in the research study were sent by e-mail to the entire twenty-three member Family Service Team (FST) of the HSC. The HSC FST is comprised of twelve RCDs and eleven FSWs. RCDs are full or part–time permanent staff who provide a range of services within large geographic regions including heavily populated areas, while FSWs are contract staff who are contracted and provide services within specific geographic areas. The Family Services Program includes: direct support services, education and support to hospitals, care facilities and other institutions, support for local community development and support to the HSC’s national advocacy efforts (Huntington Society of Canada, 2014). The only individual excluded from participation was the researcher, who was in a dual role of being both the primary researcher as well as a contracted FSW with the HSC. The researcher’s position as a FSW provided a privileged position to conduct the research as the researcher could be regarded as both a trusted insider knowledgeable about HD and as a person who understands the position, expectations, responsibilities and challenges experienced by members of the HSC FST.

The research was initially proposed to the HSC’s Director of Family Services and Community Development on January 16, 2013 who provided support and informal approval on February 7, 2013. The research was later further endorsed by the Chief Executive Officer and
Executive Director of the HSC on March 6, 2013. An internal HSC e-mail system was utilized to advise the HSC FST of the planned research through an Informal E-mail Advising of the Research (Appendix A). The University of the Fraser Valley (UFV) Research Ethics Board issued a Certificate of Human Research Ethics Board Approval (Appendix B) on March 5, 2013. The HSC FST members were then each sent a Recruitment E-Mail (Appendix C) as well as an electronic Letter of Informed Consent (Appendix D). Those who agreed to participate were asked to sign and return the consent form to the researcher electronically, by fax, or by mail. Those who agreed to participate and who had returned signed Letters of Informed Consent, were finally sent a Participant Interview Guide (Appendix E) which contained further information on the research as well as a list of questions the researcher would be asking the participants during scheduled telephone interviews. Participants were asked to consent to audio-recording of the telephone interviews and a secure coding system was created to ensure confidentiality and data security. Telephone interviews were determined to be the only viable means of conducting the research, due to the geographical challenges of participants living and working across Canada. All audio recordings of the interviews were coded and contained no identifying data. All research data and recordings are being retained for five years as per UFV ethics requirements with hard-copy documents being kept in a locking filing cabinet. Electronic data will be retained on a password protected computer.

Demographic information was gathered from participants at the beginning of the interviews, prior to transitioning into the semi-structured qualitative portion of the interviews. Participant Interview Guides (Appendix E) consisting of 13 open-ended questions were utilized to maintain focus and uniformity through the interviews, as well as to make data analysis more manageable. Interview lengths ranged from 30 to 75 minutes, with the majority of the interviews
completed in 60 minutes or less. The researcher invited additional information from participants who were also encouraged to share stories and examples from their practice experience; the researcher probed for further information and invited further elaboration as deemed appropriate by the researcher to clarify, expand upon, or to capture further information not discussed elsewhere in the interview. Finally, prior to concluding the interviews, participants were invited to add any additional information they might wish to contribute.

Data analysis was completed through a careful review of the information provided by participants, including reviewing audio-recordings and coding notes taken during the course of the interviews. Following the creation of a long compiled list of the participants’ responses to the numbered questions, responses were assigned to categories based on frequency (Dudley, 2011) as well as through carefully watching for themes to emerge from the responses (Grinnell, Gabor, & Unrau, 2010) which then led to the creation of new categories. While some data may have not been captured through this means, categorizing the data contributed to provide a useful overview of the responses most often received. Theme analysis consisted of examining the data collected throughout interviews by reviewing notes taken during the interviews and through re-listening to the recorded interviews in an effort to identify patterns or themes evident in multiple interviews (Dudley, 2011). Theme analysis also proved to be a useful strategy for evaluating the data received as research participants shared many of their own practice experiences in their work with clients.

**Ethical Considerations**

The research participants recruited were all university educated professionals who work with clients and families affected by HD as a regular part of their positions. Participants were
reminded their involvement in the research was entirely voluntary and that there was no pressure, requirement or expectation from the HSC for them to participate. Participants were not compensated in any way for their participation, although the majority of the participants were able to schedule their telephone interviews during the course of their work days with the HSC. Participants may have had concerns regarding the confidentiality of the information they shared and may also have had concerns about the compiled information being provided to the HSC. The researcher strived to alleviate these concerns by reminding participants of both their and their clients’ confidentiality. Assurance was provided that only aggregate information would be shared. Client and participant specific information was disguised to ensure no data would be attributable to any particular client or participant. Participants were also reassured that, in the event quotes or specific examples are used, identities would be kept confidential and any potentially identifying information would be omitted or disguised within the quotes or examples used in the research report. Participants were also reminded of their right to discontinue participation at any time during the interview and of their right to have any data they have contributed to the research withdrawn until the time their individual data had been compiled with the other participants’ for aggregate data analysis.

I acknowledge being employed as a contracted FSW with the HSC creates the potential for a real or perceived conflict of interest. My role could be viewed as putting me in a position where there could be a positive bias towards the HSC and the services provided or that there may be reluctance to report any finding not seen to be favorable towards the HSC. The research was conducted as an independent investigation with no restrictions or cautions from the HSC in any regard, other than to ensure no direct research would be conducted directly with HSC service recipients. The research project was initiated on the advice and with the full support of the BC
Resource Center Director (RCD), yet the research was not commissioned by the HSC. Further mitigating any perceived conflict of interest, my focus was aimed at exploring the strengths, assets, resources, strategies and successes of clients and professional staff, rather than conducting a critical analysis of what is not working for individuals or for the HSC service delivery system.

**Study Limitations**

There were several potential limitations which may have impacted the validity of this research. The researcher’s role as a FSW with the HSC may have influenced the responses of participants who may have offered greater or less information than if the research had been conducted by an outsider. Due to the time constraints involved in conducting the research within the framework of the MSW Program at UFV, the researcher was unable to access individuals with HD or their caregivers directly in order to gain their valuable and first-hand perspectives regarding what they experience as contributing factors in maximizing the quality of life of the person with HD. While having FSWs and RCDs share their wealth of experience and knowledge of having worked with hundreds of people with HD adds a valuable perspective, without the direct voices of the people with HD and their caregivers, it is difficult to determine first-hand what those with HD feel adds value, meaning, purpose and quality to their lives and to the lives of others living with HD.

The small sample size presents another limitation to the generalizability of the research findings, while the vastness of the service delivery area of the HSC raises questions as to the availability and uniformity of services and supports and the wide variability of service delivery models used across the country. The research design was further limited as the researcher relied solely on telephone interviews with research participants; the researcher suspects through the use
of focus groups participants may have prompted and triggered further comments, suggestions and strategies research participants may not have shared without the prompting and dialogue which would have occurred had focus groups taken place.

**Findings**

The response rate from invited participants in this research was high with sixteen of twenty-two or 72% of the invited professionals participating. The HSC professional staff members who participated in the research have an average length of employment with the HSC of eight and a half years. Eleven of the sixteen respondents have been with the HSC for greater than three years, while three of the respondents have been with the HSC for greater than twenty years. As the interview data suggested, long-serving team members act as experienced mentors, guides and sources of support and information for the newer employees. Although participants were not questioned directly about their employment satisfaction with the HSC, the vast majority of respondents voluntarily spoke positively and passionately about their employment with the HSC. One participant reported:

> I feel so very fortunate to be able to work for such an incredible organization. The clients and families are such an amazing source of inspiration and the organization itself is unlike any other non-profit I have ever worked for. The connections made by the HSC between the scientific community, the professional staff and the families have served to create an incredibly strong sense of community everyone benefits from.

The employment satisfaction voluntarily expressed by HSC staff, average length of employment as well as the high response rate to this research seems to suggest HSC staff are an engaged group of professionals committed to the agency and to the clients and families whom they serve.
Through a careful analysis of the information gathered from research participants, the following findings and ten themes emerged from the research:

- Defining Quality of Life
- Self-Awareness
- Stigma
- “Self” and Huntington Disease
- Family and Huntington Disease
- Community and Geographical Variability
- A Little Help from my Friends
- Grief and Loss
- Advance Care Planning
- Consumer Directed Care

**Defining Quality of Life**

As previously mentioned, quality of life is subjective. Yet there can be little doubt the presence of a degenerative neurological disorder will impact quality of life regardless how it is defined. Engstrom and Nordeson (1995) conducted quality of life interviews with 169 individuals with a variety of neurological diagnosis and discovered the following themes among participants who were asked how they would define *quality of life*: living with a sense of affinity; being independent; living on one’s own terms and finally, feeling that life is meaningful were all seen to be defining factors in understanding or describing quality of life. Participants in my own research echoed the results found in Engstrom and Nordeson’s research with participants unanimously mentioning the importance of social connections and relationships with others as well as the need to find purpose in living. Several participants mentioned the importance of maintaining an ability to contribute in some meaningful way with many suggesting maintaining employment whenever possible, while others spoke of the importance of being in touch with spirituality and nature. Research participants were unanimous in mentioning social determinants of health as factors impacting clients’ ability to maintain the quality of their lives. Factors such
as: financial security, recreation, access to hobbies, access to health-care, employment opportunities, housing, political stability, diet and nutrition and access to the basic necessities of life were identified as major factors which allowed individuals to maintain a reasonable quality of life, despite the existence of HD in their lives.

**Self-Awareness**

Dellefield and Ferrini (2011) suggest that progressive neurocognitive decline is often even more troubling than the physical symptoms of HD. Neurological decline is manifest by impulsive and often aberrant behaviors, personality changes, psychotic or depressive symptoms, perseverative activities or thinking patterns, restlessness and anxiety, poor judgment and a lack of self-awareness (p.187). The majority of research participants suggested their clients with HD often demonstrated limited ability to self-assess their own disease progression and lack the self-awareness needed to identify appropriate supports, services and resources available which could be helpful and relevant to them through the course of their disease; these observations are consistent with the literature and are indicative of the disease’s impact on cognitive functioning, reasoning and problem solving (Banaszkiewicz et al., 2012). Several research participants reported clients may have been aware of supports available within their communities, yet the clients with HD were unable to recognize the available services would be of benefit to them. Banaszkiewicz et al. (2012) suggest the disease is perceived very differently by patients and their physicians with physicians able to see the disease progression objectively while the patient is only able to view the disease subjectively along with the perceptual limitations which are a part of the disease process. Snowden et al. (1998) reports perceptual limitations can be demonstrated by the fact that most people with HD are unaware of their involuntary movements although the movements are obvious to others. Although research participants provided a long list of
services, supports and resources available to support individuals requiring assistance, participants also reported their clients with HD often struggled without utilizing formal supports thinking their symptoms and condition did not warrant the use of supports in their circumstances. One participant commented:

Through a combination of lack of insight, trouble navigating systems and supports that are ill-equipped to meet people with HD’s needs, it is really no wonder so many people with HD don’t bother to even try to access services and supports which may in the end help support their independence and their ability to live in their own homes longer. Their lack of self-awareness is definitely the primary barrier to people with HD getting the supports they need.

The disease process which contributes to clients’ lack of self-awareness, was one of the many reasons research participants cited why individuals living with HD tend to do significantly better when they have actively involved caregivers in their lives. Caregivers can help to attend appointments, provide collateral information to healthcare providers and to find tactful and creative ways to introduce support services into their loved ones’ lives.

The literature reviewed and several of the research participants interviewed concurred, individuals with HD’s lack of self-awareness can also contribute to aggravating existing relationships, often with caregivers, family members and the friends closest to those with HD. According to participants, those with HD frequently lack the ability to fully appreciate the impact and demands their disease puts on their loved ones. Participants reported those with HD can be demanding, impulsive and unrealistic in their expectations of their caregivers, which can further strain relationships and disrupt family roles and functioning. As individuals with HD
progress through the disease process, communication can become increasingly difficult which leads to progressive dependency putting caregivers and family members at great risk of burn-out due to constantly increasing demands. One participant suggested: “given that those with HD lack the ability to self-assess, it becomes critical caregivers become skilled at establishing and maintaining boundaries and self-care strategies in order to counter their loved one’s lack of self-awareness”.

Despite the HD disease process contributing to an impaired sense of self-awareness, one research participant suggested with focused attention and support, those with HD may be able to nurture and maintain a reasonable degree of self-awareness if this skill is practiced and developed early on in the disease process. The participant suggested through education about HD and the disease’s progression, as well as through engagement with others who have HD, affected individuals might learn to better recognize their own symptoms and could share their experiences with others. The participant believed those with HD could learn strategies to minimize the effects of the disease and could develop more effective strategies to adapt and adjust as needed to the progressive symptoms expected through the advancing stages of the disease.

**Stigma**

During the course of the research study, participants were asked to reflect on both the factors that contribute to quality of life in people in the general population and to then consider the unique challenges people with HD face in maintaining a high quality of life. The vast majority of respondents suggested the stigma associated with having HD unnecessarily
contributed and further aggravated individuals with HD’s ability to live high quality lives. One respondent elaborated:

People with HD are misunderstood, judged and stereotyped based on misconceptions and often because of an intoxicated-appearing presentation. The misconceptions and ignorance about HD pushes people with HD further underground increasing isolation, apathy and depression and further preventing community and societal integration.

This quote highlights the importance of communicating and connecting with HD clients and to ensure they are supported and included as active members of their communities. Kralik et al. (2006) suggest connecting with others and sharing stories of both struggles and successes is critical in building resiliency in those living with chronic illness and disease.

Kaptein et al. (2007) allude to the impact of stigma for individuals with HD suggesting quality of life itself is socially constructed and is dependent on the beliefs fostered by the social environment regarding people with HD and of the disease itself. Several research participants cited stigma, self-consciousness and embarrassment as contributing factors which prevents clients from feeling welcome and engaging in community activities and events. Social isolation was seen to contribute and aggravate depressive symptoms which in turn was reported to leave those with HD even less likely to want to take the effort to involve themselves in their communities.

Stigma also appears to impact people with HD at a systemic level. Stigma and discrimination against those with HD is nothing new, and has been well-documented for decades, with Perry (1981) suggesting people with HD are discriminated against and are routinely denied insurance coverage, their drivers’ licenses are revoked unnecessarily, and
qualified job applicants are denied positions in favor of applicants who do not carry the mutant gene. Perry (1981) goes on to report how lay organizations are active in many countries fighting discrimination and “lobbying to change laws that discriminate unfairly against mentally and neurologically handicapped persons” (p.1100). It is interesting to note, almost thirty years later, the HSC continues to align itself with other neurological and genetic organizations to fight discrimination and to reduce the stigma individuals and families with HD still face today. In fact, on April 18, 2013 the Winnipeg Free Press (Rennie, 2013) reported Senator James Cowan, the Liberal Leader in the Senate, introduced legislation that would stop insurance companies and others from discriminating against people who are genetically susceptible to some diseases. Senator Cowan drew attention to a University of British Columbia study (Bombard et al., 2009) which revealed widespread discrimination against people at risk for HD, most often by insurance companies, family members and in social settings as well as reports of discrimination at work, doctors’ offices, and hospitals and by the different levels of government. While there are currently no laws in Canada that specifically prohibit genetic discrimination, there does appear to be broad political consensus around the issue and we can only hope, perhaps finally we are on the cusp of ending genetic discrimination as we know it in Canada. Nonetheless, until this occurs, Bombard et al. (2009) suggest even those individuals who are not yet symptomatic but who are at genetic risk, will also face the risk of discrimination simply because they have a family history of HD.

All of the research participants shared they see it as part of their role to educate, inform and raise the profile and awareness of HD in the communities in which they work. Participants expressed how they believe they are well-positioned to help support the development of an HD community and feel they are able to educate others on how they too can raise public awareness
of the disease. Research participants were unanimous in their suggestion that individuals with HD do far better when they are connected to others in their communities and as one participant suggested, “HD is a dreadful disease… something nobody has to deal with on their own”.

“Self” and Huntington Disease

As one participant stated, HD does not present on a “blank canvas” but rather colours, shades, and is added to who a person already is prior to being diagnosed with the disease. Individuals with HD may be “new to HD” with no previous knowledge of the condition and no known family history, or the individual may be the most recent family member to receive a positive diagnosis in a long line of family members impacted by the disease. Regardless of the family history, individuals bring with them their own personalities, character, identity, values, strengths, attitudes, biases, coping strategies and history amongst other things when they receive their diagnosis. Research participants unanimously agreed an individual’s attitude impacts how they will approach and deal with their HD and their attitude will also influence how they deal with the world around them both before, and after HD. One participant suggested:

Those who were friendly, positive, optimistic and well-connected socially before being diagnosed tend to do better when compared to those who were negative, critical and pessimistic or who tended to isolate before being diagnosed. I suppose positive people are just easier to be around and attract others to them, while negative and grumpy people tend to push others away.

According to several research participants, social workers and other professionals working with individuals with HD would be well advised to consider each client holistically; clients’ whole person health needs to be treated and supported, not just the physical, but also the social,
emotional, psychological, spiritual and relational well-being, as all are connected and all contribute to clients’ over-all wellbeing and to overall quality of life.

In discussion with research participants, it became clear a diagnosis of HD could have very different meanings depending on the individual receiving the diagnosis. Participants reported individuals did not have a “typical” response to learning of their diagnosis and reactions were as varied as the people being treated. Participants further shared how the traits, characteristics and attitudes of those diagnosed with HD continued beyond diagnosis with those who had been resilient, connected, optimistic and who had positive attitudes before diagnosis appeared to take these traits with them post diagnosis.

Participants suggested people with HD, not unlike those without the disease, are the product of their pasts, their upbringings, their cumulative life experiences and, of course, their genetics. Participants said clients add HD to who they are, but it doesn’t have to necessarily define who they are. It was suggested those individuals who are successful at incorporating HD into their lives and then live their lives as fully and completely as possible, despite the HD, fare far better than those who allow the disease to define who the individuals are as people. It was unanimous among participants, HD makes optimizing the quality of one’s life more difficult, especially in the later stages, however it has also been unanimous, the attitude the individuals diagnosed with HD bring with them into HD has a significant impact on the quality of life that individuals ultimately lead. One research participant suggested:

When times get tough we all have to dig deep and draw on inner resources to pull ourselves back up, those with HD have to dig that much deeper and try that much harder to find the strength and spirit to carry on.
Family and Huntington Disease

Research participants reported HD significantly impacts the quality of life of those affected, but the disease also impacts caregivers and family members who themselves may be at risk of having inherited the mutant gene. As a genetically inherited disease, HD is very much a family matter. Therefore, it is understandable a great deal of research has been conducted on assessing and evaluating the quality of life of caregivers and family members as well (Aubeeluck, 2005; Aubeeluck, & Buchanan, 2007; Helder et al., 2002; Roscoe et al., 2009). Individuals at risk for HD carry their families with them on their journey; they may carry the legacy and memories with them of a parent, sibling(s), or extended relative(s) who had lived or who is living with HD and they may make comparisons backwards and forwards based on their experience of what they have seen with their loved one’s experiences and quality of life with HD.

Kaptein et al. (2007) report “quality of life research is increasingly being directed at both the impact of the social environment on the quality of life of chronically ill individuals, and at the toll the illness exacts on the quality of life of close family members” (p.793). A participant in Carrozzi and Tulsky’s (2012) research offered the position that “people at risk for HD see themselves fast-forward into that time space” (p.223). Individuals learn from each other, both how to be with HD themselves, as well as how to care for another with HD. Individuals and families might choose to detach from others, choosing to live in relative isolation alone and oblivious to the support, assistance, resources, and community which could be of assistance to the individual or family with HD. In some cases, individuals and families just may not realize there is any other way to be with HD.
The majority of participants shared stories of clients and families who never speak openly about HD. These participants reported that clients have told them that being diagnosed, treated and supported is “nothing more than a wishful dream” because they want their loved ones to take a proactive approach. Several participants reported that their clients may have learned from their immediate and extended families how to be with HD, both through seeing them as living examples and through having them openly sharing lessons learned along the way. Participants further shared how families teach their loved ones to welcome the HSC as well as both formal and informal supports into their lives. Participants also shared how, in contrast, other families do the opposite and deny the disease and the need for supports, as if to accept a diagnosis would be akin to cursing both themselves and by extension their family with a disease they would rather deny.

**Community and Geographical Variability**

Canada is a vast country with varied geography, weather systems and access to services and supports. While there are several federally funded programs and services such as Old Age Security and Canada Pension Plan Disability Benefits which are largely uniform across our country, other programs and services are offered through provincial and municipal governments as well as through health authorities, non-profit, local and community organizations. Participants reported access to provincial, regional and municipal programs and services are highly variable with some clients with HD having to travel hours or even days to access supports, while others have both informal and formal supports and services readily available. Participants suggested services and supports across Canada are as variable as the needs of individuals who make up the HD community.
Research participants shared stories of successes and challenges accessing services with no definitive conclusion suggesting whether rural or urban residents with HD are better served. According to participants, people with HD in urban centers generally appear to have increased access to primary care, specialists and to multi-disciplinary teams and also benefit from easier access to transportation, yet the cost of living in urban centers can often be much higher than those who live in rural areas. People with HD in rural areas were reported by participants to have less access to standardized care than their urban counterparts and often have significant challenges with transportation, distances, and lack of formal services. People with HD in rural communities were, however, reported to be resilient, resourceful and were able to draw upon informal support systems to receive personalized care suited to their unique needs. One participant working in a rural area reported:

People with HD in rural areas struggle to meet even their basic needs and attending the HD clinic or seeing specialists is extremely difficult. The planning, coordination, distances, weather and expenses are all challenges and many folks who would definitely benefit from attending the clinic simply aren’t able to access the care that others in urban areas are able to enjoy.

This quote highlights that people with HD in rural areas were reported to be more reliant on others for transportation, for community connections and to assist with coordinating the long, arduous trips to HD medical clinics. This is also dependent on whether the person with HD was fortunate enough to have a clinic accessible to them. Research participants’ comments also demonstrated how social determinants of health can impact clients’ and families’ ability to access supports and services and to how well they are able to integrate into the communities in which they live.
A Little Help from my Friends

Without exception, research participants agreed the quality of life of individuals with HD is enhanced by the presence and involvement of an active family, friend or other informal caregiver(s) engaged in the person’s life. Caregivers (who in this paper, refers to unpaid, informal caregiving including family and friends) were reported to be “critical” to the well-being or the person with HD. As one participant illustrated, seeming to capture the spirit and thoughts of all of the research participants: “people with partners, hands down, have a better quality of life than those who are on their own”. Participants suggested individuals living with HD who have the active involvement of caregiver, know they have a consistent person in their lives they can trust and who will be a solid, stable secure force in their lives. Research participants further suggested caregivers help open doors to resources, act as advocates and aid in communicating their loved ones care needs, preferences and history especially in the event communicating has become compromised as a result of the disease progression. Caregivers provide collateral information to health care teams, which is especially valuable when the person with HD’s self-reports are inaccurate or incomplete. While this research did not focus on differentiating between paid and unpaid caregivers, the vast majority of caregivers referred to by research participants were unpaid, and were most often close family members, typically the person with HD’s partner or spouse.

While the person with HD benefits from the active involvement of their caregiver, several research participants suggested that caring for the person with HD can become “suffocating”. With a considerable risk for co-dependent relationships developing, it is critical respite for the caregiver is available and that delicate balance is found and maintained for parties giving and receiving care. O’Connor and McCabe (2012) report satisfaction with social relationships,
whether marital or general social relationships, is a consistent predictor of quality of life both for individuals suffering from neurological disorders as well as for their caregivers. The majority of participants stressed the importance of professionals providing support, not only to the person with HD, but also to the caregivers and family members involved in providing care. People with HD are parts of broader systems, families and communities; participants suggested that the HSC professional staff recognizes the importance and value of the inter-connected systems supporting people with HD and strive to nurture people with HD and those in the community around them.

**Grief and Loss**

In addition to the disease process of HD (characterized by avoidance, denial and lack of self-awareness, as frequently mentioned by research participants), individuals with HD also are prone to suffer from depression and apathy. This can make initiating activities difficult. For example, several participants suggested clients with HD might neglect personal care or housekeeping tasks in part due to the difficulty posed in performing the tasks, but also because initiating the task might be more than the person with HD is able to overcome. One participant suggested “for many, the perceived benefit of completing a task is simply not worth the effort”. Research participants reported people with HD often wait “until it is too late” to access services and supports that could be of benefit to them and further tend to have a delayed response to the need to prepare in advance for their disease progression. One respondent suggested caregivers may further contribute to this delay advising:

> Caregivers often want to protect their loved ones from what the caregiver perceives to be the un-pleasant task of preparing for one’s own disease progression or their own demise and doesn’t push for supports their family member might benefit from.
This quote suggests while caregivers may have more accurate perceptions of their loved ones needs and deficits, their desire to protect their loved ones from the acknowledgement of the disease progression, might result in some individuals and caregivers choosing not to access supports which may be beneficial to the individual with HD.

More than half of the research participants shared how they believed their clients’ cumulative losses and unresolved grief also contributed to individuals having trouble initiating tasks and new activities. Anger, isolation and the inability to maintain employment and participate in activities formerly enjoyed were all seen to contribute to depression, apathy and a loss of optimism and self-confidence. Participants suggested clients’ reduced independence, changes in self-identity and loss of vision for a meaningful future all contribute to a reduced ability and desire to initiate tasks or to develop new connections. One participant suggested “the progression of HD is like a slow death… all hope is gone.”

**Advance Care Planning**

Research participants stressed the importance of ACP for all people, but especially for those with HD. One participant suggested “while most of us talk about being prepared in case of what *might* happen, for people with HD it’s more a matter of being prepared for what *will* happen”.

Advance care planning (ACP) is an on-going process of reflection and communication in which a person who has decision-making capacity makes decisions regarding future health and/or personal care in the event that he or she becomes incapable of consenting to or refusing treatment or other care (Heyland et al., 2013, p.1).
The respondents unanimously reported people with HD do not make full use of available ACP tools such as representation agreements, power of attorney documents, advance directives and wills. These legal and logistical issues were identified as an area needing further attention. According to participants, education and encouragement from the professionals involved would better prepare both people with HD and their caregivers to complete ACP in a timely manner. Participants suggested they believed barriers to participants completing ACP included the belief that the process was too complicated, was better done very near the end of life, was seen as unpleasant or even morbid. One participant suggested “when you are living with HD, there are just always other more pressing and urgent issues needing attention”.

Participants suggested it is important ACP be introduced early and often. Planning for the future should be presented in a way that reminds people it ensures their voice will be heard when they are no longer able to speak for themselves. The majority of participants also suggested ACP needs to be “normalized” and that people with HD shouldn’t be singled out; it is equally important caregivers and all family members complete ACP tools. One participant suggested social workers involved with families impacted with HD need to reconcile the fact that “while we may feel obliged to ensure our clients maintain hope, this also ignores the reality of the disease progression and the need for planning in advance for residential care, life-support issues and end of life planning”. Another participant shared they were aware of a large family having an “ACP party” where a professional facilitator assisted many family members to complete tools appropriate for each of their circumstances in a safe, non-threatening and reportedly “fun” environment. Several respondents suggested those with HD are not unlike the rest of the population in that they tend to want to avoid tasks perceived as being unpleasant, however, planning in advance ensures clients’ voices are heard, their wishes are respected and
that they ultimately maintain at least some degree of control over not only the quality of their remaining life, but also might take comfort knowing that they also retain at least some degree of control over the quality of their deaths as well.

**Consumer Directed Care**

Research participants reported existing home support services and available community services were often not appropriate to meet the needs of individuals with HD. Services available were reported by participants to frequently provide a standardized model where services appear to be designed more for the efficiency and benefit of the system, rather than to meet the unique needs of the clients being served. Participants shared how those individuals who were thriving had pieced together a web or “smorgasbord” of services rather than expecting the services of one agency or organization being adequate to meet all of the client’s needs.

While not widely available across Canada, and often mired in bureaucracy in areas where it is available, formal systems of consumer directed care were suggested to be a potential option to optimize the quality of life for some individuals with HD. Consumer directed care puts financial resources in the hands of the individual or family needing care rather than in the hands of the government and has been demonstrated to provide more personalized client centered care than that provided through a standardized case management assigned model. In an Australian study on the effectiveness of new approaches providing personal support, Fisher, Parker and Purcal (2009) concluded:

It is evident, future models of support and housing provision for persons with disabilities, traditionally needing 24-hour support, should irrespective of the housing setting or level of support needed, focus on an individual approach. This can facilitate mobility and
flexibility as needs change and it can provide options for integrating informal, formal and generic support. The research shows this approach is also most likely to meet the goals of supported living policy in terms of human rights, quality of life and independent living (p.331).

While consumer directed care in its current model is not well-suited for many people with HD who lack the ability to direct their own care, perhaps a future incarnation of this model of care will provide greater options and choices for those needing greater flexibility than our current models allow.

Research participants spoke of what they regarded to be the generally inappropriate setting of extended care or residential care facilities for those with HD. With HD typically presenting mid-life, several participants suggested younger individuals with HD don’t fit well within care facilities, as those with HD are often much younger than the rest of the facility population. Although not widely accessed in Canada at present, consumer directed care provides some potential for those with HD requiring increased care to explore the possibility of remaining living in the community with personalized supports. Several participants suggested social workers and those employed by the HSC FST are in a position to advocate for those with HD and to lobby for increased access to programs, services and funding which would allow more personalized and appropriate care to those who have the support network and the desire to remain at home.

**Implication for Practice and Policy**

Professionals employed by the HSC as well as other professionals engaged in the care and support of individuals with HD would benefit from considering the results of the current
research. A definite need has been demonstrated to actively work towards increasing education regarding HD, in an effort to reduce the stigma associated with the disease and to promote community involvement and inclusion for those living with HD. There is reluctance and in many cases an inability by individuals with HD to recognize the need for and to accept available supports and services, especially on a preventative basis. Yet, it is also clear those who do engage with the broader HD community and who do accept both formal and informal services and supports benefit tremendously as a result. Professionals would do well to continue to explore creative, non-threatening and effective ways to introduce supports and services to people with HD as early as possible in their disease progression.

The HD community including those families directly affected by HD, professionals working with the community and the HD scientific community need to continue to work together to conduct further research into what contributes to the optimization of the quality of life of those impacted by HD. Further research needs to be carried out to explore the strengths, strategies and resiliency that contribute to the quality of life of individuals who manage to thrive despite the very real challenges living with HD entails.

These research findings may support professional staff by encouraging them to examine both the literature and their own beliefs around what contributes to improved quality of life for clients. The research will assist professionals to maximize their role as helpers by supporting them to share with their clients the supports, services and strategies identified that have proven to be useful to others and to invite their clients to consider utilizing the strategies themselves. Ultimately, it is the researchers hope successful strategies to optimize the quality of clients’ lives will become apparent and well recognized and that all those who could benefit from the research
will have access so as to optimize the quality of their own lives - whatever that might mean to them.

**Dissemination**

This report will also be offered for inclusion on the HSC website which is accessed by those in the HD community from around the world. The completed report will also be offered to all research participants who may further share the findings with staff, clients and families with whom the research participants are involved. The researcher’s position as a contracted worker with the HSC also affords the opportunity to present the findings at smaller, regional events such as the three annual retreats for people with HD held across Canada. The research results may also be useful to other individuals and professionals involved with other neurodegenerative disorders and depending on timing, availability and interest, the research findings may also be offered to other interested organizations.

Dissemination of the research results and the generalizability of the findings will also require further careful ethical consideration. While ideas, examples and strategies others have found helpful may be tempting to adopt for oneself, each individual, family system and community is different with unique strengths, challenges and needs. It is hoped the research findings will be useful to individuals, families and professionals within the HD community. However, each person considering using information contained within the final report would need to carefully weigh the pros and cons of applying the information or strategies to their own specific situation.
Recommendations for Future Research

Current literature offers a variety of assessment tools to measure quality of life and health-related quality of life, yet there is limited research offering few specific suggestions on how to improve the quality of life of individuals living with HD. The research conducted as reported in this paper only begins to explore the specific factors that influence and enhance the quality of life of those living with HD, and a great deal of further research is needed in this area.

This research has invited the perspectives of key staff working with the HSC from across Canada who generously offered their wisdom, experience and perspective, yet the perspectives and direct voices of those with HD and those providing care remain unheard. It is the researcher’s hope that further research will be conducted seeking to identify successes, celebrate the spirit and identify the critical components and variables necessary to create and maintain high quality lives for people living with the progressive neuro-degenerative symptoms of HD.

Research participants were unanimous in their agreement that those with HD who have the care and support of a loved one do considerably better in managing the course of their disease when compared to those who progress through the course of the disease on their own. Further research differentiating formal and informal care as well as exploring the differences in both types of support systems would be beneficial to better understand the pros and cons and benefits and challenges of both types of care.
Conclusion

HD is a disease that challenges individuals to discover and maintain meaning, purpose, and quality of life. This is despite the reality they have been delivered a neuro-degenerative sentence that will not only impact them, but will also stretch their family and social support networks, and will leave their children with a 50% risk of acquiring the disease themselves. Individuals’ and families’ pre-symptomatic functioning and ways of being will provide them with a foundation on which they will build their response to the threat that is HD, while also contributing to the quality of life they are able to maintain while simultaneously fighting their battle.

The current research has identified the importance of establishing and maintaining relationships and connections with others, and to being connected within the community in which one lives. Resources, supports and information is available to individuals and families living with HD through both formal and informal means, although through a lack of awareness, or perhaps through denial and fear, many do not connect with relevant and available support systems until far too late to realize the full benefits. Ten of the sixteen participants specifically mentioned the HSC as a critical resource for those living with HD. More than half of the participants reported HDRCs were also extremely helpful for those with HD who have access to the multi-disciplinary specialized clinics. Several participants also suggested the HSC’s Family Service Team provides support, education, connections and referrals to those living with, and to those at risk of HD, and striving to better prepare clients and families for the journey ahead and encourage early engagement with the HD community and with appropriate supports.
The HSC FST participating in the research shared how they are moved, touched and inspired by the courage, dignity and grace with which many people with HD face their diagnosis and symptoms. In spite of the tragic way HD impacts people physically, emotionally and cognitively, participants shared exceptional stories of how individuals lived lives filled with meaning, purpose and quality regardless of the challenges. The HD community has countless living examples of spirit, conviction and courage and would do well to find ways to increase the opportunity to share these stories and demonstrate that despite HD, those affected are able to live lives they can be proud of and can provide legacies of courage, resilience and bravery for those who follow.
References


doi:10.1080/10410230902804133


Appendix A

Informal E-Mail Advising of Research

February 13, 2013

INVITATION TO ALL RCDs AND FSWs

Greetings RCDs and FSWs,

In the next week or two I will be inviting each of you to consider joining me in an exploration of what factors contribute to Optimizing the Quality of Life of Individuals with Huntington Disease.

In brief, I will be commencing my major research project for my MSW which will entail conducting telephone interviews with as many RCDs and FSWs that are willing to participate. I’m of course aiming for 100%, although of course you are under no obligation to participate. I anticipate each telephone interview will take 30-45 minutes and will examine your observations, experiences and thoughts on how individuals with HD can optimize the quality of their lives.

The interviews will be semi-structured, yet I assure you, there will be ample opportunity for you to share stories of success, triumph and the strength of spirit so often seen amongst our clients with HD.

Please look for a more formal invitation to participate in the coming weeks and please consider participating and adding your voice and the experiences of your clients to this research.

I anticipate I will be ready to schedule the telephone interviews for very end of February and early March and I will be most flexible in accommodating your schedules and our various time zones.

I am happy to answer any questions you might have in anticipation of the research and I welcome your early expressions of interest willingness to participate.

Thanks for considering this request and please look for more information in the near future.

Warm regards,

Randy Goossen, FSW – Fraser Valley, British Columbia

E-mail: rgoossen@huntingtonsociety.ca

Voicemail: 604 851-0225
# Appendix B

UFV Ethic Board Certificate

---

## Certificate of Human Research Ethics Board Approval

<table>
<thead>
<tr>
<th>Contact Person</th>
<th>Department</th>
<th>Protocol #</th>
</tr>
</thead>
<tbody>
<tr>
<td>Randy Gooszen</td>
<td>Social Work</td>
<td>549</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Co-investigators</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adrienne Chan</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Title of Project</th>
</tr>
</thead>
<tbody>
<tr>
<td>Optimizing the Quality of Life of individuals Living with Huntington Disease</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sponsor/Funding Agency</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Institution(s) where research will be carried out</th>
</tr>
</thead>
<tbody>
<tr>
<td>University Of The Fraser Valley</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Review Date</th>
<th>Approval Date</th>
<th>Approval Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>14-Feb-13</td>
<td>05-Mar-13</td>
<td>5-Mar-13 - 04-Mar-14</td>
</tr>
</tbody>
</table>

**Certification:**

The protocol describing the above-named project has been reviewed by the UFV Human Research Ethics Board and the procedures were found to be in compliance with accepted guidelines for ethical research.

---

Kathy Keiver, Chair, Human Research Ethics Board

*NOTE: This Certificate of Approval is valid for the above noted term, provided there is no change in the procedures or criteria given. If the project will go beyond the approval term noted above, an extension of approval must be requested.*
March 6, 2013

Hi Everyone,

At long last, I have received final ethics approval for my MSW research titled Optimizing the Quality of Life of Individuals with Huntington Disease.

As HSC's Resource Center Directors and Family Service Workers, I want to invite each of you to consider participating in a 30-45 minute tele-interview to discuss in your experience what contributes to some people with HD thriving while others struggle.

Please find attached more detailed information on my research as well as a Letter of Informed Consent for those of you who are willing to participate. While you are of course under no obligation to participate, I would be absolutely thrilled if over the course of the next month or so if I could interview all of the RCDs and FSWs employed by the HSC.

If you are willing to participate, please return the signed consent form to me ASAP and then we can schedule a time for an interview at your convenience. After receiving your signed consent, I will send you a list of questions I will be asking during the interview to allow you time to consider your responses in advance of the interview.

Thanks again for considering this request and for being willing to share your knowledge, wisdom and experience.

Warm regards,

Randy Goossen, RSW
Appendix D

Letter of Consent

University of the Fraser Valley
33844 King Road
Abbotsford, BC V2S 7M8
604 504-7441
March 5, 2013

Optimizing the Quality of Life of Individuals Living with Huntington Disease

Letter of Informed Consent

My name is Randy Goossen RSW and I am a student in the Masters of Social Work program at the University of the Fraser Valley as well as a contracted Family Service Worker with the Huntington Society of Canada. As Family Service Workers or as Resource Center Directors with the Huntington Society of Canada, you have been identified by the researcher as key informants with valuable perspectives into how individuals with Huntington Disease (HD) can live life to the fullest.

Purpose/Objectives of Study

In this study I will explore the factors that key informants believe contribute to how some individuals with HD are able to live with a high quality of life despite being faced with the challenges of living with a degenerative neurological disorder.

Procedures Involved in the Research

Due to participants living and working in a variety of areas across Canada, the researcher will rely on telephone interviews for the vast majority of data collection. The researcher will schedule times to talk with all participants so as to fit within participants’ schedules. It is expected the telephone interviews will take 30 – 45 minutes to complete. The researcher will be seeking your permission to audio-record and take notes during interviews to ensure information shared is accurately captured. In the event you prefer not to be audio recorded, your participation will still be welcomed, although the researcher will need to take notes in the absence of audio recording to ensure data obtained during the interview is accurately captured.
Telephone interviews will begin with a brief preamble, definitions will be provided for key terms and the researcher will ask for demographic information before beginning the main part of the interview which will consist of capturing observational and qualitative answers to 12-15 questions.

**Potential Harm, Risk or Discomfort**

It is unlikely there will be any harm, risk or discomfort involved in participating in this research. Participants are free to withdraw from the research at any time and are also not obligated to answer any questions they choose not to answer. Participants are welcome to take breaks or discontinue the interview at any time by simply notifying the researcher of their desire.

**Potential Benefits**

The research will contribute considerable experiential knowledge to the existing knowledge base on how to optimize the quality of life for individuals living with HD. Service and care trends may be identified both for those directly impacted by HD and for professionals providing services. Professionals advocating on client’s behalf will have increased knowledge and awareness on the services, supports, strategies and approaches that are most effective in improving the quality of life for those living with HD. The research may identify gaps in services and supports and may assist individuals affected by HD and the professionals involved in their care to advocate and design supports and services to better meet the needs of those living with HD. It is hoped the research will both highlight what is currently working to optimize the quality of life for those living with HD and will also demonstrate where there are areas needing further research and attention to further improve the lives of those living with HD.

**Confidentiality**

Interview notes and audio recordings will only be reviewed by the researcher who will compile and analyze data and will then shred the notes and delete the audio recordings once the data analysis is complete. All data will be secured in a locked filing cabinet and electronic data will be stored on a password protected computer. Participants’ names will be coded and names will not be used within the report nor during the dissemination of the results.

**Participation**

Participants are free to withdraw from the research at any time and are not obligated to answer any questions they choose not to answer. Participants can also withdraw from the research after the interview with all data obtained during their interviews being destroyed providing the participant’s withdrawal occurs before the data is compiled with other participants’ data.

**Research Results**

The completed research will generate a final paper which will be submitted as a graduation requirement for the MSW program at UFV. The research will also be shared with the Huntington Society of Canada for use as they see appropriate and will be offered by the researcher for consideration for presentation at future HSC National Conferences. An electronic copy of the completed report will also be offered to all research participants who advise the researcher they would like a completed copy. The final report will be completed by May 2014.
Questions

Please note the ethics of this project have been reviewed and have been approved by the UFV Human Research Ethics Board. If you have any questions regarding this research, please feel free to contact the researcher by phone at 604 557-2205 or by e-mail at randy.goossen@student.ufv.ca. Any concerns about the research may be directed to Mr. Brad Whittaker, Director, Research Services and Industry Liaison at UFV at 604 557-4044 or via e-mail at brad.whittaker@ufv.ca.

CONSENT

By signing below I agree to participate in the study, Optimizing the Quality of Life of Individuals Living with Huntington Disease.

I have read the information presented in this Letter of Informed Consent regarding the research being conducted by Randy Goossen and Dr. Adrienne Chan at the University of the Fraser Valley. I am aware I have the opportunity to ask questions regarding my involvement in this research and to receive any additional details I might require.

I understand I have the right to withdraw from the study at any time and that my confidentiality and anonymity will be preserved. Any concerns may be brought to the researcher, Randy Goossen or Brad Whittaker, Director of Research Services at 604 557-4044 or via e-mail at brad.whittaker@ufv.ca.

Please circle one option to indicate your preference below:

a) I agree to be audio-recorded and am aware the researcher will be taking notes during the telephone interview.

b) I do not want to be audio recorded, but I am aware and agreeable that the researcher will be taking notes during the telephone interview.

Name (please print)  ____________________________________________

Signature  ____________________________________________

Date  ____________________________________________

Once signed, please retain a copy of the consent form for your records and then either fax a copy of the signed consent to Randy Goossen at 604 556-5010 or scan and e-mail the consent to randy.goossen@student.ufv.ca or mail the consent to:

Randy Goossen

#20-32925 George Ferguson Way

Abbotsford, BC

V2S 6Z9

Thank you again for agreeing to participate in this research.
**Definitions**

I am well aware as professional staff with the Huntington Society of Canada, you are well-informed on Huntington Disease, but in order to ensure I am accurately capturing the information you are providing during your interview, I want to provide a few basic definitions:

**Huntington Disease** – I am referring to the genetically inherited neurodegenerative disorder characterized by motor, cognitive and psychiatric disturbances (Hocaglu, M.B., Gaffan, E.A. & Ho, A.K. (2011).

**Optimization** – I am referring to the act, process, or methodology of making something as fully perfect, functional, or effective as possible (Merriam Webster, 2011 Retrieved February 3, 2013 from http://www.merriam-webster.com/dictionary/optimization).

**Person with HD** - I am referring to an individual who has tested positive for the mutant gene and who may or may not be symptomatic.

**Quality of life** - I am referring to the World Health Organization definition of quality of life as being an individual’s view of their own place in life, in terms of their cultural and value system and in relation to their own goals, expectations, standards and occupations (Sehanovic, Dostovic, Smajilovic and Avdibegovic, 2011).
Appendix E

Participant Interview Guide

PARTICIPANT #
PARTICIPANT INTERVIEW GUIDE

Title: Examining the Factors that Contribute to the Optimization of the Quality of Life for Individuals Living with Huntington Disease.

Preamble

Thank you for your willingness to participate in this research.

I have received your signed informed letter of consent and want to remind you of a few things before we begin the interview.

I want to remind you that while you have agreed to participate in this research, you also have the right to discontinue your participation at any time during the interview and may request your responses be withdrawn from inclusion in this research up until the time the raw data is compiled for data analysis.

Before beginning the qualitative portion of the interview, I would like to begin by asking a few demographic questions in an effort to gain a better understanding of the professional staff providing support to people with HD in Canada.

I also want to remind you that the information you share, including your demographic data will be held in strictest confidence and will only be shared as aggregate data once all participant demographics and interviews have been completed and compiled.

Audio-recording

A) In your letter of informed consent you agreed to have our interview audio-recorded. I will advise you when the audio-recorder begins and ends and want to assure you again that all audio-recordings will be deleted upon completion of data analysis. I will also be taking notes throughout our interview.

or

B) In your letter of informed consent you did not agree to have our interview audio-recorded. I will therefore only be taking notes during our interview. I want to assure you all notes will be kept in a locked filing cabinet until the data compilation and data analysis is complete at which time all notes will be shredded.
I expect this interview will take 30 – 45 minutes to complete. Please let me know if you would like to take a break or discontinue this interview at any point and please feel free to advise me if there are any questions contained in the interview that you prefer not to answer. I can also discontinue the audio-recording at any time upon your request.

**Definitions**

With your letter of consent I also sent you a short list of definitions I thought would be helpful for you to be familiar with before our discussion. Do you have any questions regarding the definitions I sent you where the terms *Huntington Disease, Person with HD, Quality of life* and *Optimization* were defined?

Do you have any other questions before we begin the interview?

I will now begin the interview and will start the audiotape and/or begin taking notes.

**Begin Audiotape or Begin taking Notes**

“This research is being conducted as a requirement of the Masters Degree of Social Work Program at the University of the Fraser Valley”. The purpose of this research is to examine the factors that you, as a professional employed by the Huntington Society of Canada, believe contributes to the optimization of the quality of life for individuals living with Huntington Disease”.

**Demographics**

“I would like to begin by asking a few demographic questions in an effort to gain a better understanding of the professional staff providing support to people with HD in Canada”

1. Sex
2. Age
3. Degree(s) obtained
4. Your position with the HSC
5. Length of employment with the HSC
6. Are you employed FT or PT by the HSC

**Questions**

1. Not thinking specifically of HD but rather of people in general, what do you believe are the factors that contribute to people’s quality of life?

2. Do you believe HD creates additional challenges in establishing and maintaining a high quality of life? If so, can you describe the additional challenges?
3. In what way do you believe the progression of HD can impact a person with HD’s quality of life? Please explain.

4. Are there strategies you have seen people with HD employ to successfully optimize the quality of their lives despite the progression of the disease? Can you describe these strategies?

5. Do you believe there are strategies people with HD can employ early in their disease progression that are likely to increase the quality of their lives at later stages of the disease progression? If so, can you discuss these strategies?

6. What formal supports and services do you believe can contribute to the optimization of individuals with HD’s quality of life?

7. What informal supports and services do you believe can contribute to the optimization of individuals with HD’s quality of life?

8. Are there supports or services you believe are under-utilized by people with HD? What are these services? How might they be helpful? Are there barriers accessing these services?

9. How important do you believe it is for people with HD to have a family or caregiver’s support to assist them to optimize the quality of their lives?

10. To what degree do you believe a person with HD’s attitude toward their HD impacts their quality of life?

11. Do you believe people with HD fully utilize formal advance care planning tools such as representation agreements and power of attorney documents? If not, what do you think could be done to increase the use of these tools?

12. In what ways do you believe in your position as a professional employed by the HSC, you can assist people with HD to optimize the quality of their lives?

13. Is there anything else you would like to add in regards to optimizing the quality of life of people living with HD?
Thank you again for agreeing to participate in this research.

Conclude Audio-recording and/or Discontinue Taking Notes